Wunderlich Syndrome: A Life-Threatening Condition

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Wunderlich syndrome is an uncommon condition of spontaneous renal hemorrhage in the subcapsular and perirenal space, of atraumatic etiology in the kidney, with the potential to spread to the retroperitoneal region beyond the perirenal fascias. Its clinical manifestations usually include Lenk's triad, namely, acute flank pain, flank mass, and hemodynamic instability but seen in less than a quarter of patients. The clinical symptoms vary and are nonspecific which vary depending on the causative underlying renal pathology. Tumor bleeding of benign and malignant renal neoplasms is the most common cause of this syndrome, followed by vascular disorders and renal cystic diseases. Here we report the two cases of wunderlich syndrome due to renal angiomyolipoma bleed which were managed successfully.

Keywords: *Angioembolization, Angiomyolipoma, Renal hemorrahage, Wunderlich.*

Wunderlich syndrome (WS), a rare condition, is defined as a spontaneous, non-traumatic renal hemorrhage confined to the subcapsular and perirenal space.[1] Though various etiologies have been associated with this condition, angiomyolipoma of the kidney is the most common neoplasm associated with spontaneous renal hemorrhage. Bonet described the first case report in 1700 and Wunderlich in 1856, where he reported a syndrome characterized by a spontaneous renal hemorrhage with intrarenal, perinephric, or subcapsular space.[1] Lenk triad is present in 20% of the cases of WS and consists of acute flank pain, flank mass, and hypovolemic shock. However, the clinical

symptoms vary and are nonspecific.[2] A Computed Tomography is considered the gold standard in establishing the diagnosis and often elucidates the underlying etiology attributing to this condition.[3] The patient can be managed either conservatively, or by selective Arterioembolization and/or by Surgical interventions.

Case 1

A 35-year-old female presented to Emergency department with the complaint of left flank pain for 2 weeks, off and on, severity has increased in past few hours, associated with nausea and vomiting. She gave no history of trauma, hematuria, fever, rigors or chills. She was a known case of Hypothyroidism under medications otherwise no associated other medical illness or history of drug intake. On examination her vitals were stable, with left flank tenderness and left renal angle tenderness positive. Her blood profile report was normal except slight increase in total leukocyte count. Her ultrasound report showed evidence of a large ill-defined fatty mass of 13.4 cm x5 cm in size seen closely lying in the posteroinferior part of left kidney causing displacement of kidney upward. Her CECT abdomen showed fibrofatty proliferation and soft tissue strandings in Left perinephric space giving mass like

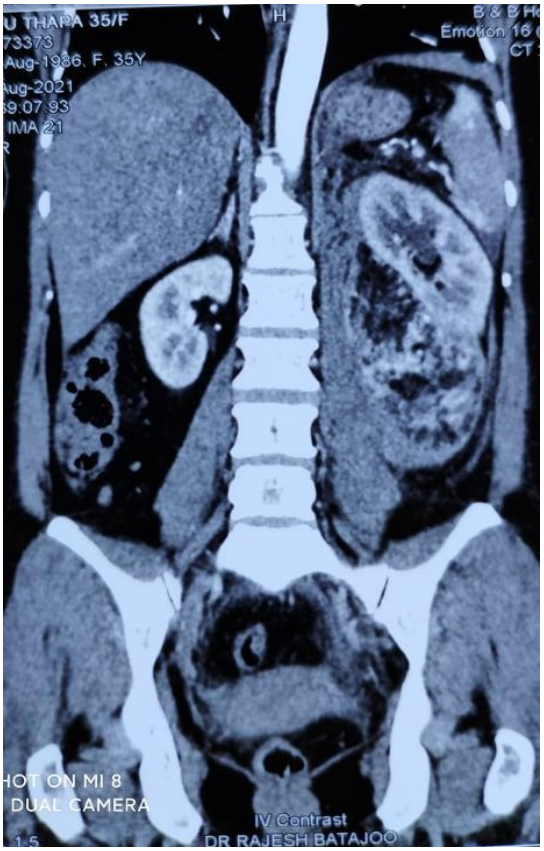

Figure 1: CECT abdomen showing Rupture Angiomyolipoma

appearance measuring ~18x11x10 cm in size in medial & inferior region encasing renal vessels and pelvis & displacing ureter anteriorly & kidney superiorly with minimal extravasation of contrast in inferior perinephric space suggestive of ruptured Angiomyolipoma (**Figure 1**).

Patient was kept under conservative management, but later she became hemodynamically unstable with drop in Hb/PCV value and needed blood transfusion. Patient underwent selective Renal Angioembolization (**Figure 2**) with the use of absolute alcohol and gel foam by the interventional Radiologist. Patient was discharged and was on regular follow

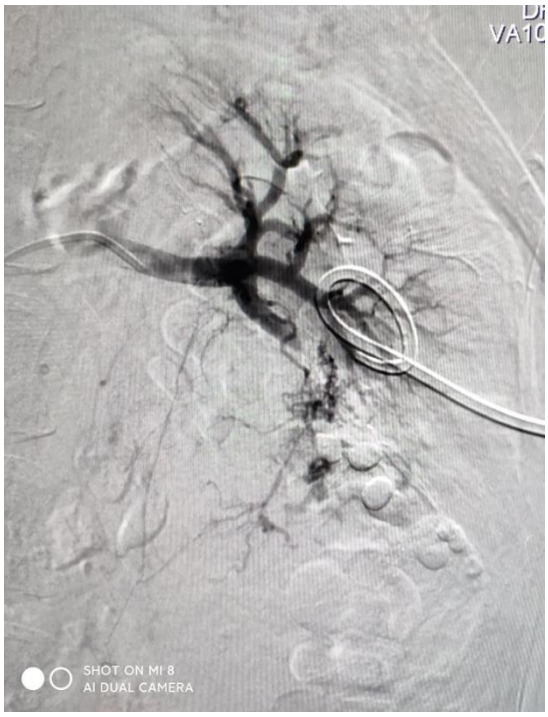

Figure 2: Selective Renal Arterio-embolization shown

up every 3 months and CT was done. Even After 6 months, there was no significant decrease in the size of the mass, and patient had persistent complaints of left flank pain.

Patient was planned for partial Nephrectomy but ended with total nephrectomy since kidney was not salvageable intraoperatively with massive bleeding from the dissection planes. On gross analysis, the mass was made up of a large hematoma enclosed within the perinephric space and a ruptured yellowish mass arising from the lower pole of the left kidney. Microscopic examination revealed that the lesion arising from the lower pole of kidney was composed of triphasic components- islands of mature adipose tissue, myoid spindle cells and dysmorphic thick blood vessels in a hemorrhagic background. The postoperative period was uneventful and the patient was discharged on ninth postoperative day. With 6 months completed after treatment, she is doing well.

Case 2

A 33-year-old female presented to out patient department for the definitive management of ruptured renal angiomyolipoma, for which she was earlier managed in some other center where she had presented with severe left flank pain and was in state of hemorrhagic shock. At our center on presentation, her vitals were stable with normal blood parameters. Her ultrasound report showed well defined, heterogeneous lesion in the mid-pole of the left kidney with minimal peripheral vascularity, likely angiomyolipoma with intramural hematoma. CT abdomen showed a large, lobulated well defined complex heterogeneously enhancing, fibrofatty mass with fat density area and non-enhancing hyperdense areas within the interpolar region of the left kidney extending to the left perinephric space, suggestive of ruptured Angiomyolipoma. Patient was planned for selective Renal Angioembolization. Post angioembolization she developed post embolization syndrome characterized by

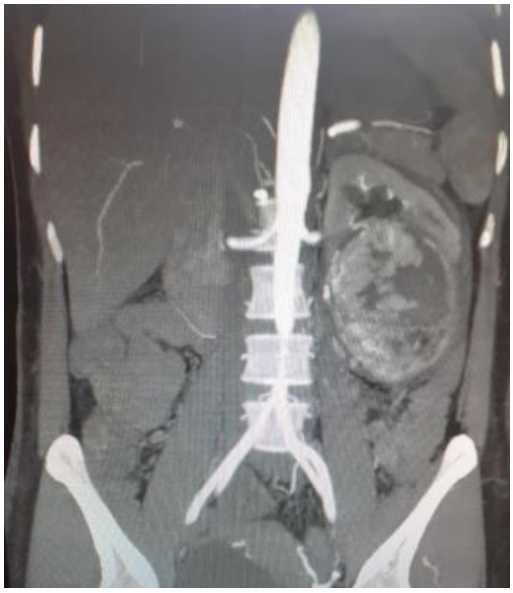

Figure 3: Left Rupture Angiomyolipoma

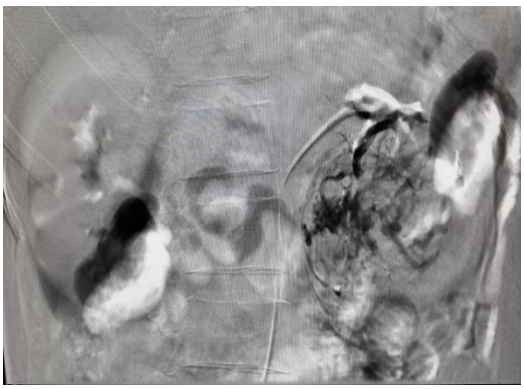

Figure 4: Selective Renal Arterio-embolization

fever, flank pain and raised leukocyte counts. She also had drop in the Hb level for which blood transfusion done. Patient was discharged on the 7th Post-operative day. Her repeat CT image after 3 months showed significant reduction in the size of the mass.

Discussion

Wunderlich Syndrome or spontaneous hemorrhage from kidney was first observed by Bonet in 1700 and first clinically described as 'spontaneous renal capsule apoplexy' by Carl Reinhold August Wunderlich in 1856.[1] The term Wunderlich Syndrome was first used by Coenen in 1910. This is a rare clinical phenomenon with only about 250 such cases reported since 2003.[2] It is depicted as spontaneous bleeding of kidney into perirenal and sub capsular spaces following trauma.

Renal neoplasm's accounts for 60-65% of spontaneous renal haemorrhage, the angiomyolipoma being the most common benign neoplasm.[3] Classically, Wunderlich Syndrome is described by the presence of Lenk's triad which constitutes acute flank/lumbar pain, palpable tender mass, and features of active internal bleed like hypotension, tachycardia and anemia.[4] Clinically however, this triad is rarely seen and presents with abdominal pain (67%), hematuria (40%) and hypovolemic shock (26.5%). The other symptoms being nausea, vomiting, low grade fever and anemia. A Computed Tomography is considered the gold standard in establishing the diagnosis and often elucidates the underlying etiology attributing to this condition.[3] Magnetic Resonance Imaging is a valuable alternative if other radiological investigations are equivocal.

In treating Wunderlich Syndrome, achieving hemodynamic stability is of

foremost importance. In the pursuit of achieving this, endovascular and/or surgical interventions play a major role. Although, Trans arterial embolization has emerged as initial choice of treatment in spontaneous renal hemorrhage conventional exploratory surgery holds its domain especially in patients with unidentified source of hemorrhage after radiological evaluation.[5] Preoperative contralateral renal status assessment is a necessity in all cases as one may land up doing total nephrectomy due to unforeseen circumstances as it was in our first case. Patients, who have been treated conservatively, needs follow up at three months intervals so as to know the status of the hematoma.[6] One must also be ready to face a malignancy which may have been missed in the initial radiological investigations especially, the renal cell carcinoma being the most common malignant neoplasm.[3]

Conclusion

Cornerstones in management of a case of Wunderlich Syndrome includes high index of suspicion, prompt resuscitative measures, focused investigations, identifying the underlying cause, embolization and/or surgery. Still, the choice and timing of the treatment modality needs to be tailored based on the resources available and should be individualized to achieve optimum results. In order to obtain better insight into this rare condition all diagnosed cases however trivial should be published. The low incidence of Wunderlich Syndrome could be attributed to the varied presentation, lack of suspicion and moreover the diagnostic difficulties encountered. All these factors contribute to faulty diagnosis and under reporting of the Wunderlich Syndrome.

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